Alveolar Soft Part Sarcoma of the Retroperitoneum

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ALVEOLAR SOFT PART SARCOMA, an unusual neoplasm of uncertain histogenesis,[1] has only rarely been reported in the retroperitoneal area. Smetana's[2] comprehensive study[2] recorded only four such cases and the average age of the patients was 25 years. In the case here reported the patient was a 73-year-old woman and the first clinical symptoms were those of partial obstruction high in the intestine associated with severe secondary anemia.

REPORT OF A CASE

A 73-year-old white woman was admitted to Santa Barbara General Hospital on January 2, 1957, with complaint of progressive weakness, anemia, nausea and vomiting of six months' duration. There was a positive reaction for occult blood in the stool. Upper gastrointestinal roentgenograms showed medial displacement and partial obstruction of the second portion of the duodenum by a large mass in the right retroperitoneal area. Pronounced hydronephrosis on the right was demonstrated by an intravenous pyelogram, and retrograde pyelography showed decided distortion and displacement of the right upper ureter (Figure 1).

At laparotomy a large retroperitoneal tumor involving the right kidney and extending over the second and third portion of the duodenum was observed. It was considered nonresectable. The mass, approximately 20 cm. in diameter, appeared to be extrarenal and attached to the right psoas muscle with displacement of the right ureter and moderate secondary hydronephrosis of the right kidney. The external surface of the tumor appeared quite vascular and the cut surface of tissue obtained for biopsy appeared firm, gray and slimy. A palliative anterior gastroenterostomy was performed to by-pass the obstruction.

Postoperatively the patient received irradiation (tissue dose 3,000 r) to tumor mass in the right upper quadrant, with only minimal improvement. Weakness and anemia progressed, due to chronic loss of blood associated with ulcerative tumor extension in the duodenum. The patient died seven months after operation and approximately a year after onset of symptoms.

Pathologist's Report

At autopsy the retroperitoneal mass was observed to extend to the hepatic flexure of the colon, to the second and third portions of the duodenum and to the lower pole of the right kidney (Figure 2). Dissected from contiguous structures, the mass was a lobulated, ovoid tumor measuring 30 x 25 x 15 cm. Cut surfaces were pale ivory-white. The tumor filled the upper right retroperitoneum, producing a 15 x 6 cm. ulcerated tumor mass in the third portion of the duodenum. The posterior aspect of the tumor was attached to the right psoas muscle. Multiple distant metastastic lesions were present in the liver, pleura and lungs.

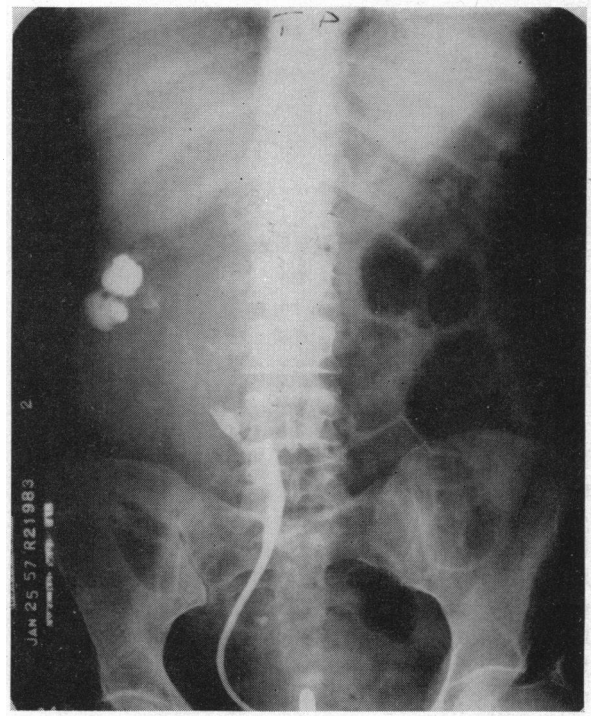

Figure 1.—Retrograde pyelogram showing distortion of right upper ureter due to retroperitoneal alveolar soft part sarcoma.

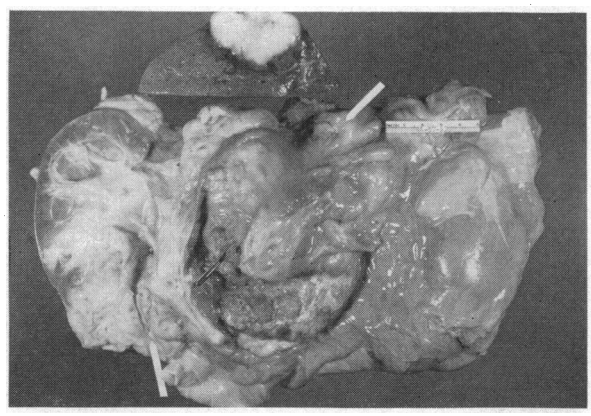

Figure 2.—Retroperitoneal alveolar soft part sarcoma, showing in situ relationship to right kidney and ulcerated duodenal tumor extension. (Probe in ampulla of Vater. Markers at pylorus and right ureter. Liver wedge with metastasis above.)

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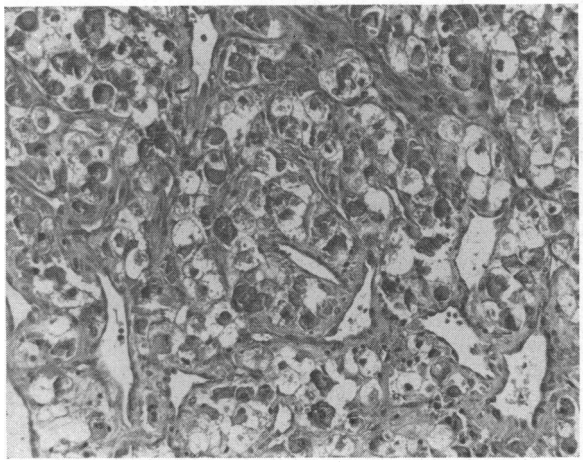

Figure 3.—Primary retroperitoneal alveolar soft part sarcoma (×100) showing characteristic alveolar cell grouping and prominent sinusoidal vascular pattern.

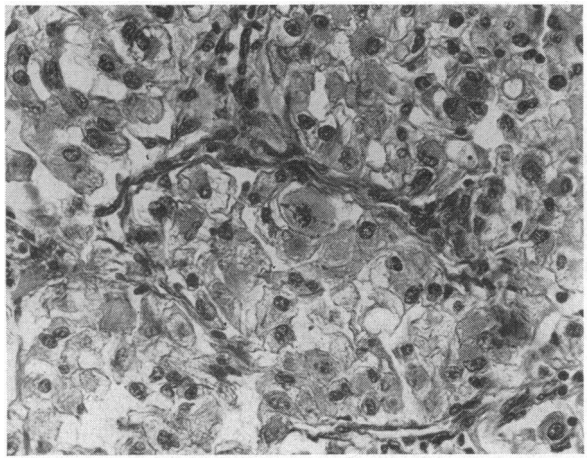

Figure 4.—Gomori trichrome (×400). Pulmonary metastasis of alveolar soft part sarcoma, showing preservation of original growth pattern, with scattered mitoses, one of which is visible centrally.

Microscopically, the tumor consisted of irregular, rounded groups of large cells surrounded by capillary vascular spaces which often appeared compressed, but occasionally were dilated in sinusoidal fashion, producing an organoid or "endocrine" type of arrangement (Figure 3).

Individual tumor cells generally appeared polyhedral or spheroidal in outline. They ranged from 15 to 70 micra in diameter and had an abundant eosinophilic granular or homogeneous ground-glass cytoplasm which often appeared vacuolated but contained scanty sudanophilic material and no demonstrable chromaffin granules. Cell nuclei were rounded to ovoid with vesicular or reticulated chromatin, usually showing a single, slightly acidophilic nucleolus. Mitoses were rare in the original surgical material, but averaged one to three per 10 high power microscopic fields in the postirradiation autopsy specimen (Figure 4).

SUMMARY

A case of alveolar soft part sarcoma of the retroperitoneum in an elderly woman is reported. Autopsy findings included ulcerative invasion of the duodenum and distant visceral metastasis following irradiation therapy.

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Brucellosis—A Case Study

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THE CHRONIC FORM of brucellosis may present many subjective symptoms which are difficult to distinguish from those of psychoneurosis—symptoms such as excessive fatigability, myalgia, low-grade fever, depression, insomnia, headache, anorexia and loss of weight. In the acute form of the disease symptoms are more objective. The onset of acute brucellosis is variable, but usually is initiated by fever of the rising "septic" type with morning remissions. Associated with the fever are a slow pulse, neutropenia, diarrhea, headache, depression, muscular aching, fatigue and shifting joint pain. When fever is of the undulant type, febrile episodes persist for ten to twelve days and are followed by periods when the patient is afebrile and asymptomatic. In most cases there are no such undulations and the patient remains ill for two weeks to ten months. In the differential diagnosis of brucellosis, consideration should be given to typhoid fever, Q fever, miliary tuberculosis, chronic recurring infectious mononucleosis, pulmonary or systemic coccidioidomycosis, rheumatoid arthritis, and Boeck's sarcoid.

The clinical diagnosis of brucellosis, therefore, must be substantiated by laboratory diagnosis. Of the many laboratory procedures available, the only absolutely diagnostic criterion is a positive blood culture. If routine blood cultural methods are used, the chance of identifying and isolating the organism

From the Services of Dr. A. G. Bower, Chief Physician, Communicable Disease Unit of the Los Angeles County General Hospital.

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